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Bouveret Syndrome, A rare cause of Gastric Outlet Obstruction

Clinical Presentation

A 64-year-old female had an acute admission with complaints of coffee ground vomiting, acute onset abdominal pain and a two-week history of nausea, epigastric pain and indigestion.

She had a past surgical history of previously failed elective laparoscopic gallbladder operation.

Her vital observations were within normal limits and the blood tests performed on admission were found unremarkable.

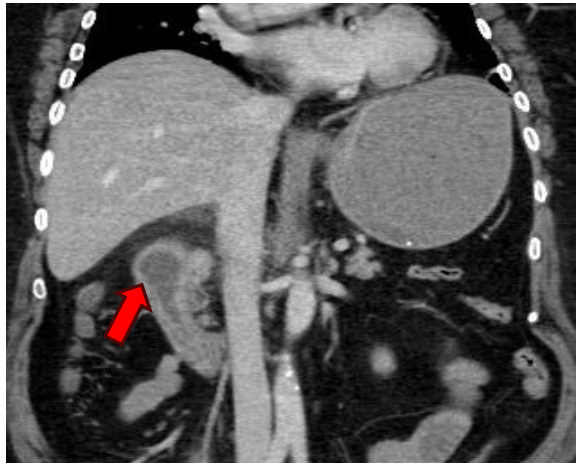
What Investigations did she have?

A CT abdomen with portal venous contrast was performed on the same day which showed:

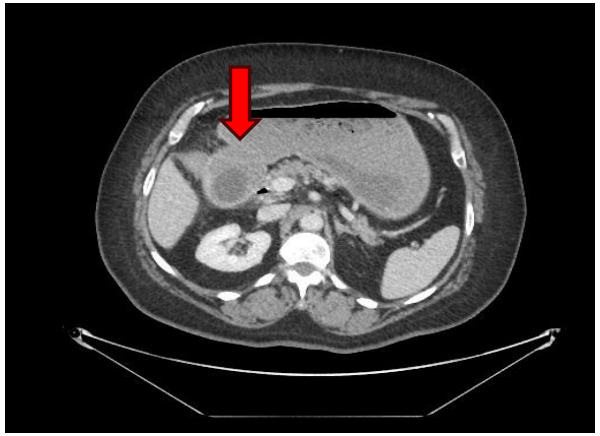
"Irregular gall bladder walls with tiny air pockets within the lumen and surrounding walls with inflammatory changes in adjacent fat. Gallbladder lumen was seen communicating with pyloric canal with a lesion resembling a collection in close contact with duodenum and gastric outlet, causing the outlet obstruction. Pneumobilia was also noted. "

But a second review by a different radiologist concluded that this lesion was in fact a hypodense non-calcified large iso-dense gall bladder calculus residing in the distal stomach. This was in line with patient's past history of gall stones which could not be removed due to failed a laparoscopic surgery, however, this was unusual as the usual path of stone migration is distal bowel and, in this case, it had migrated upwards.

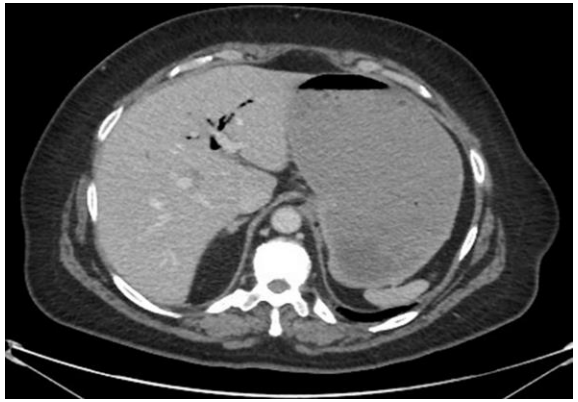
A diagnosis of Bouveret Syndrome was labelled based on all these features.



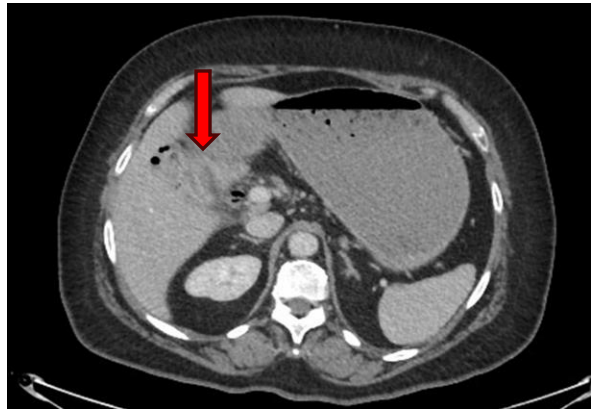
Arrow pointing towards the hypodense gall stone in the D1



Arrow pointing towards non-calcified gall stone in the distal stomach.



Pneumobilia and distended stomach



Contracted GB with pockets of gas and Pneumobilia.

Next plan of action

As it was now confirmed that a gall stone was responsible for her symptoms,patient underwent an OesophagoGastroDuodenoscopy (OGD) which confirmed a 3-4 cm stone in the pylorus with an attempt for extraction but all in vain.

She was later considered a candidate for Laparotomy which showed a cholecystoduodenal fistula with a gall stone residing in D1 and causing obstruction. A gastrostomy was performed and the stone was finally extracted.

What is Bouveret Syndrome?

Bouveret syndrome, named after L. Bouveret, a French physician, who published two comprehensive case reports in 1896 of this condition, is characterised by a rare form of Gastric Outlet obstruction caused by a gall stone in the Pylorus or duodenum that has migrated through a bilioenteric fistula which could be either a Cholecystoduodenal fistula (seen in 60% of the cases) or a Choledochoduodenal/cholecystogastric fistula each comprising about 5% of cases.

It is the most infrequent variant of Gall stone ileus, accounting only 0.3 to 5% of cholelithiasis complications.

Risk factors for this syndrome include past history of cholelithiasis, gall stones greater than 2cm to 8cm and age older than 60 years. Of all the patients who suffer, about 43% to 68% have had a past history of biliary colic, jaundice and acute cholecystitis.

It can also very rarely occur secondary to gall bladder malignancy.

The main pathophysiology is chronic inflammation and pressure necrosis, which together with mechanical pressure applied by gallstones on the gallbladder itself and bowel wall, could result in a tear of the gallbladder and enteric wall lying in approximation to each other.

Mortality

Although a rare entity, the mortality rates for this condition are high mounting up to 12% to 30% possibly due to increased age at presentation, prolonged biliary disease and other comorbidities.

Radiological Findings

Radiological features that we need to look for are

Rigler's Triad consisting of

- Gastric Outlet Obstruction
- Ectopic Gall stone
- Pneumobilia

These features could be detected either on a CT, MRCP, US or Endoscopy, however the sensitivity for finding these features is the greatest (75% to 78%) using a CT making it a more practical. Despite high sensitivity of the CT scan, its ability to detect iso-dense stones poses a main limitation.

MRCP provides the most accurate assessment of the fistulous tracts however it's not the first line of investigation in most clinical situations.

US also helps in identifying gallstones and pneumobilia but its sensitivity remains only 11% as features of the condition such as gastric distention can limit the view.

OGD has dual benefits of helping in diagnosing and treating (removal of the stone) but the chances of visualising ectopic stones are much lower compared to the CT scan (about 69%).

Treatment

- OGD is generally used as a first line for stone retrieval due to age group and co-morbidities associated with Bouverets syndrome and also it is less invasive compared to other treatments , however its only successful in about 10% of the cases.
- The mainstay of treatment is Surgery, for which the most commonly performed procedures are enterolithotomy and gastrotomy. The success rate of surgical treatment approaches 90%.

Take home message

1. A non-calcified gall stone can be easily missed on a CT scan as most of the cholesterol gall stones are either hypo or Iso-dense, therefore it is important to rule out gall stone as a cause of ileus specially in the presence of other features such as Pneumobilia.
2. Diagnosis of Bouverets Syndrome highly relies on Radiological findings and if missed could lead to an increase in the morbidity and mortality.

References

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